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METHODS

Management challenge: A case of aniridia and aphakia with secondary glaucoma in sclerokeratoplasty

Shreya Thatte[1*], Komal Jaiswal[2] and Radhika Maheshwari[3]

[1]Prof and Head, Department of Ophthalmology, Sri Aurobindo Institute of Medical Sciences and PG Institute, Indore, Madhya Pradesh, India
[2]Junior Resident 3rd year, Department of Ophthalmology, Sri Aurobindo Institute of Medical Sciences and PG institute, Indore, Madhya Pradesh, India
[3]Junior Resident 3rd year, Department of Ophthalmology, Sri Aurobindo Institute of Medical Sciences and PG institute, Indore, Madhya Pradesh, India

***Correspondence:**
Shreya Thatte,
shreyathatte@gmail.com

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A patient presented to us with post-infectious keratitis and a large anterior staphyloma in the left eye with vision as low as only the perception of light and accurately determining the projection of rays. A B-scan showed no abnormality in the posterior segment. Sclerokeratoplasty with an 11-mm graft with aniridia and aphakia was done. On 4 weeks of follow-up, intraocular pressure (IOP) was 39 mmHg on applanation tonometry, for which medical management was tried using a combination regime that was insufficient, following which a series of management approaches like trabeculectomy and Ahmed glaucoma valve (AGV) implant was done (in superotemporal quadrant). Unfortunately, recurrent exposures of the AGV tube were encountered, which were managed using different graft materials one after the other but were unsuccessful. Ultimately, AGV explantation and reinsertion at another site (in inferotemporal quadrant) was tried, which led to salvaging the eye with well controlled IOP, clear graft, and good visual function (best corrected visual acuity of 6/12p with +12.00/+1.50 × 90°). The purpose of this article was to highlight that despite multiple complications encountered in a single case, a careful sequential approach, good surgical skills, and a never giving up attitude lead to excellent results.

Keywords: sclerokeratoplasty, secondary glaucoma, trabeculectomy, ahmed glaucoma valve, conjunctival autograft, buccal mucosal graft, amniotic membrane graft, BCVA (best corrected visual acuity)

1. Introduction

The protrusion or outpouching of uveal tissue from any weak point of the eye is termed as staphyloma. Untreated or nonhealing infective keratitis often leads to the formation of anterior staphylomas. Routinely used treatment options for anterior staphyloma include dural patch graft, penetrating keratoplasty, and sclerokeratoplasty. (1–3) Despite advances in surgical techniques in large graft surgeries, the limbal support is lost, and there is a possibility of sudden expulsion of lens iris diaphragm during surgery, leading to various complications like postoperative chronic inflammation of ocular surface and anterior segment, aniridia, aphakia, graft rejection, significant astigmatism, cystoid macular edema, and secondary glaucoma. (4) The chances of intraocular pressure (IOP) rise is more in Aniridia. In addition, the chances of IOP rise after penetrating keratoplasty (PK) are higher in the aphakics. (5) The chances of graft' failure are at spike if immediate and efficient interventions for IOP control post keratoplasty are not performed. Moreover, these interventions (whether medical or surgical) may interfere with the graft's viability and survival in a negative manner (6). Our case was a rare and difficult one where multiple complications were encountered in a single case, that is, a large corneal opacity with dense iridocorneal adhesions, aniridia, aphakia,

post-PK glaucoma, failed medical management, failed trabeculectomy, and recurrent exposures of the Ahmed glaucoma valve (AGV) tube. This case report highlights that all these complications can be efficiently managed in a sequential manner in order to salvage the eye with good visual function.

2. Case details

A 42-year-old female presented with post-infectious keratitis anterior staphyloma in the left eye involving almost the whole of the cornea with visual acuity for light perception and accurate PR (**Figure 1**). A B-scan showed a normal posterior segment. Sclerokeratoplasty with an 11-mm graft was done. Intraoperatively, while separating the dense adhesion of the iris with cornea, there was a sudden, very high positive pressure, which led to the extrusion of lens iris diaphragm. After that, the anterior automated vitrectomy graft was sutured with 10-0 monofilament nylon. Postoperatively, she was started on the routine PK regime (systemic and topical steroids and antibiotics, topical antiglaucoma drugs, topical lubricants, and topical cycloplegics). In the immediate postoperative phase, the patient did well with unaided visual acuity of FC 1 ft, a clear graft with few DM folds, aniridia, aphakia, and a normal fundus with clear intervening media (**Figure 2**).

On 4 weeks of follow-up, the patient complained of mild pain in the affected eye with headache. Her best corrected visual acuity (BCVA) was 1/60, and her IOP with AT was 39 mmHg (with topical Timolol of 0.5%). On slit lamp examination, corneal edema and epithelial bullae were seen. The patient was managed for raised IOP with oral Acetazolamide of 250 mg TDS and a combination of topical antiglaucoma drugs, which were found to be insufficient to control the IOP. Hence, a trabeculectomy was done.

Two weeks post-trabeculectomy, the IOP ranged between 40 and 50 mmHg despite oral and topical antiglaucoma drugs. The patient was planned for an AGV implant in the left eye during the sixth postoperative week. The AGV implant was performed in the superotemporal quadrant under LA (**Figure 3**). In first postoperative week after AGV, IOP lowered to 18–20 mmHg and the graft remained clear. The IOP further lowered up to 8–10 mmHg in subsequent week. Two weeks after AGV implant, the exposure part of AGV tube was observed. A conjunctival autograft (CAG) was used to cover it. After 18 days of CAG implant, the AVG tube was exposed again. This time, a buccal mucosal graft was used, and the patient was started on oral doxycycline at 100 mg BD. After 3 weeks, again a larger area of exposure of tube was seen, and the valve was also seen to be migrating temporally and anteriorly toward the limbus. This time, a dry amniotic membrane was applied to cover the valve (**Figure 4**). After 2 months, the graft was clear, unaided visual acuity was 1/60, and BCVA was 6/12p with +14.00/+0.75 × 100°, IOP on AT was found to be 7 mmHg, but the AGV had to be explanted due to recurrent exposure. It was reimplanted in the inferior quadrant (**Figure 5**).

3. Results

After 8 weeks of AGV implant in the inferotemporal quadrant, IOP was well controlled, AGV was well covered with good bleb over it, a clear graft (**Figure 6**), and a normal fundus with a BCVA of 6/12p with +12.00/+1.50 × 90°.

4. Discussion and conclusion

The most dreaded culprit for graft failure and irreversible vision loss post-PK surgery is the development of secondary glaucoma (1, 2). PK in cases of adherent leucoma, anterior staphyloma, aphakia (7), mechanical collapse, or damage of the trabecular meshwork or aniridia (8) are more commonly associated with postoperative IOP rise and secondary

FIGURE 1 | Post infectious keratitis anterior staphyloma at presentation.

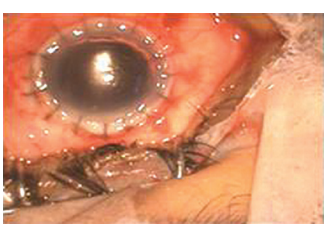

FIGURE 2 | Immediate post-op photograph with clear graft.

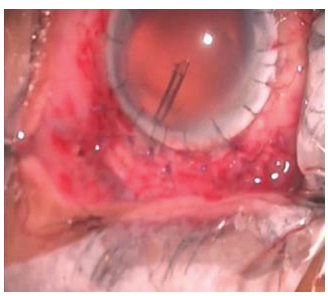

FIGURE 3 | AGV implanted in superotemporal quadrant.

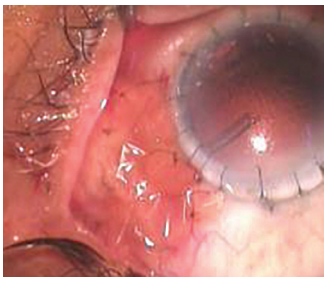

FIGURE 4 | AGV covered with dry amniotic membrane.

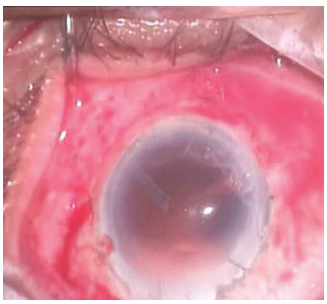

FIGURE 5 | AGV reimplantation done successfully in inferotemporal.

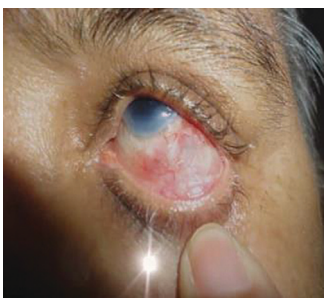

FIGURE 6 | Eight week post AGV implantation in inferotemporal.

glaucoma development. In this regard, our case was many-fold challenging because all these glaucoma precipitating factors were present in a single case in the form of dense iridocorneal adhesions, aphakia, and Aniridia. Moreover, endothelial cell loss is also common after PK, if there is a rise in IOP. (2) This makes the graft's survival very difficult. Therefore, early diagnosis and management of post-PK IOP rise can lead to preservation of the graft and aid in optimal visual function. Various treatment options discussed in the literature for such cases include medical management using topical beta-adrenergic blockers, oral carbonic anhydrase inhibitors, topical prostaglandin analogues, topical miotics in various concentrations, and topical alpha-2-adrenergic agonists. Other options include laser iridoplasty or laser trabeculoplasty or the implantation of glaucoma drainage devices (which can be valved like the Ahmed valve, or valveless like the Molteno implant) (1, 2, 8, 9) Valved devices (e.g., Ahmed glaucoma valve) have the advantage that it can be easily inserted and chances of ocular hypotony are less. (3) But implant exposure is a common complication noted in 2–7% of eyes in different literature, more common in eyes with previous ocular surgeries (7, 10, 11) and in inferonasal quadrant. In this case, after the failure of medical management and trabeculectomy in controlling IOP, an AGV implant was inserted in superotemporal quadrant, but tube exposure was observed, which was very essential to cover in order to avoid the risk of developing ocular inflammation, hypotony, phthisis, and late endophthalmitis. The use of various materials (like sclera, dura, pericardium, buccal mucosa, and amniotic membrane) has been enlisted since time immemorial to cover the exposed part. We tried conjunctival autograft, buccal mucosal graft, and amniotic membrane graft one after the other, but all these measures were unsuccessful and the grafts were not well taken up, may be because ocular surface in the superotemporal quadrant was already compromised due to chronic infectious keratitis and perforation. Finally, AGV was explanted and implanted again in the inferotemporal quadrant, where the conjunctiva was healthy, leading to successful results in terms of well-controlled IOP, a clear graft, a and good visual outcome (BCVA 6/12p).

Hence, dealing with such complicated cases requires advanced knowledge, appropriate timing, good surgical skills, and patience.

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